CASE REPORT

Propriospinal myoclonus: is it always psychogenic?

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SUMMARY

Propriospinal myoclonus (PSM) is a rare type of movement disorder in which the myoclonic jerks are present or intensified in supine position. Arising from a thoracic or abdominal generator, the movements propagate rostrocaudally as per the spinospinal conduction velocity. Routine neuroimaging and electrophysiology being normal in most instances, it is frequently interpreted as psychogenic. We present a case of PSM which will throw ample light on the pathophysiology, approach and dilemmas in treatment.

BACKGROUND

Myoclonus is a brief involuntary shock-like movement due to muscle contraction (positive myoclonus) or inhibition of contraction (negative myoclonus).¹ It has been variously classified according to the clinical features, aetiology and anatomic origin. Among different forms, propriospinal myoclonus (PSM) is rare and very few cases have been described in literature. Organic as well as psychogenic aetiology have been proposed in previous case reports.^{2–5} Although the disease is notorious for being unresponsive to treatment, drugs such as clonazepam, valproate and zonisamide have been proven to be useful. We describe a case of PSM who responded to clonazepam after 1 week of treatment.

CASE PRESENTATION

A 45-year-old woman presented to our department with jerky movement of the trunk and limbs for 6 months. According to the patient, the movements started after few hours of a scorpion bite to her right forearm. The movements were appearing only in supine or semireclining posture but were absent in sitting straight, standing or walking. They were sufficiently intense to cause shaking of the whole body. For the muscle pain and anxiety because of the abnormal movements, she was unable to relax on bed. They were persisting through out the day but most frequent between 20:00 and 22:00 when the patient was retiring to bed and they were completely absent during sleep (as reported by relatives). There was no loss of consciousness, bladder or bowel incontinence, behavioural changes, cranial nerve palsy, weakness or numbness in any limb or previous history of any major medical or surgical illness. Family history was unremarkable and there was no history of addiction.

General examination did not reveal any abnormality. Higher mental function and cranial nerve examination were normal. Nutrition, tone, power and reflexes were intact in all four limbs and

plantar response was flexor. Sensory, cerebellar and other system examinations were normal.

On examination, the patient was having involuntary, jerky, brief, shock-like movements on supine and semireclining position and they were disappearing as soon as she started to walk or even stand up. They were involving the trunk as well as the limb, symmetric, but slightly more lateralised to left. They were forceful enough to lift the body off the couch. The movements were neither being aggravated in presence of care takers nor subsided on diverting her attention, a feature suggestive of psychogenic movement disorder (video 1).

INVESTIGATIONS

Complete blood count, renal functions, liver functions, serum electrolytes, random blood sugar, thyroid profile and serum vitamin B₁₂ levels were within normal limits. EEG recording showed only the jerk artefacts but there were no epileptiform discharges preceding the movements (video 2). MRI of the dorsal spine with screening of the whole spine was performed. It revealed atrophy of the dorsal cord without any signal changes or extramedullary flow void (figure 1). MRI of the brain in T1 and T2 axial view did not reveal any abnormality.

DIFFERENTIAL DIAGNOSIS

From the jerky shock-like movements which were completely posture related, a diagnosis of PSM was made. Suspected aetiologies were mainly spinal cord pathology, psychogenic or toxin induced (as a history of scorpion bite was temporally related to the onset of symptoms).

Figure 1 MRI (A) T1-weighted and (B) T2-weighted sagittal images of cervicothoracic spine revealed atrophy of thoracic cord (arrows).

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Video 1 The patient demonstrated involuntary, jerky, brief, shock like movements on supine and semi reclining position.

TREATMENT

The patient was treated with tablet clonazepam 1 mg twice daily. The movements gradually became lesser in intensity and subsided completely after 1 week (video 3). She was discharged with the same medication.

OUTCOME AND FOLLOW-UP

During follow-up after 1 month, there was no such movement and the patient again started to lead an independent and socially productive life. She is on regular follow-up since 3 months and there is no recurrence of symptoms.

DISCUSSION

PSM is an extremely uncommon movement disorder characterised by myoclonic jerks which arise in muscles corresponding to a specific myelomere and spread to other myelomeres rostrally as well as caudally. It was Brown *et al* who provided the first description of PSM in 1991 and till date, about 50 cases have been documented in literature. This disease usually has a male predominance, unlike the present case. The myoclonic movements are postulated to take origin from a hyperexcitable segment of spinal cord, mostly thoracic or abdominal. The wave of hyperexcitability then propagates from the generator segment

Video 3 The patient showed complete absence of involuntary movements.

rostrally in keeping with the spinospinal conduction velocity. Aetiology in previous case reports mainly included spinal cord diseases (syringomyelia, arteriovenous fistula and demyelinating lesions), drugs, toxins, malignancy and psychogenic. Some cases were termed idiopathic as no definite cause could be found out. Characteristic features of the disease includes (1) worsening or appearance on supine position; (2) involvement of abdominal muscles; (3) worsening on sleep-wake transition and (4) cord abnormalities on MRI or DTI FT of spine. Diagnosis is based on the clinical features and polymyography which demonstrates the generator muscle, extent of propagation up as well as down stream and the slowed spinal conduction velocity from the latencies of electromyography (EMG) bursts. For technical issues we could not perform multichannel EMG.

Conventional MRI protocols may not demonstrate obvious cord abnormality in which we have to rely strongly on electrophysiology to make a diagnosis of this entity. In one of the case series, diffusion tractography with fibre tracking could help in documenting the cord abnormality, but we did not go for it as we got definite evidence of cord atrophy. Because of the absence of any obvious lesion on non-contrast MRI (eg, flow void, mass lesion) and the evidence of non-epileptic jerks, a contrast image was not performed.

Video 2 EEG clip showing movement artifacts corresponding to the myoclonic jerks in absence of any cortical discharge.

It becomes at time difficult to differentiate propriospinal myoclonus from two closely resembling entities, cortical myoclonus and psychogenic myoclonus because they differ in treatment as well as outcome. Cortical myoclonus has a spike discharge on EEG preceding the EMG bursts by 10–40 ms which was absent in our case. We thought of a diagnosis of psychogenic myoclonus at admission but the diagnostic clinical features such as distractibility, variability, la belle indifference were not present.

The clinicopathological correlation of PSM is still not full-proof. In some studies majority of cases were found to have a purely psychogenic origin.⁴ Definite evidence of stressor sometimes have a confounding effect even when an organic cause is found out. Hence it is utmost essential to rule out an organic cause before labelling the disorder as psychogenic for better management.

Drug treatment does not give convincing results but there are reports of spontaneous remission. Medications such as clonazepam, zonisamide, amitriptyline are helpful in relieving symptoms. Treatment of the cause if obvious on MRI may provide remission.

Learning points

- ▶ An index of suspicion for propriospinal myoclonus is needed when the jerks have a strong postural relationship and EEG is normal.
- ▶ It can well be confused with psychogenic myoclonus leading to improper treatment and failure of remission of symptoms.
- ▶ Multichannel electromyography and diffusion tractography with fibre tracking are the cornerstone of diagnosis.
- ▶ No consensus opinion exists regarding drug treatment, treatment of cause if found can produce remission.

Contributors RV formulated the hypothesis. HNP, TPR and DR prepared the manuscript and videos.

Competing interests None.

Patient consent Obtained.

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